

# Speech suppression without aphasia after bilateral perisylvian softenings (bilateral rolandic operculum damage)

Villa G., Caltagirone C.

Clinica Neurologica, Università Cattolica del S. Cuore di Roma.

*The authors describe a patient who suffered two successive, right and left, strokes that caused bilateral rolandic operculum damage. The clinical picture was characterized by selective impairment of volitional facio-pharyngo-glossomasticatory movements with sparing of automatic and reflex motor activity (Foix-Chavany-Marie syndrome). Though completely speechless, the patient was not aphasic. This dissociation is discussed in the light of the peculiar localization of lesions evidenced by CT-scan.*

**Key-Words :** Bilateral rolandic operculum softenings — Foix-Chavany-Marie syndrome — speech suppression without aphasia — CT-scan.

## Introduction

Bilateral destruction of the rolandic operculum caused by perisylvian infarctions may result in the so-called 'bilateral anterior opercular syndrome' [9] or Foix-Chavany-Marie (FCM) syndrome [5]. This uncommon neurological disorder is characterized by the selective impairment of voluntary movements — with preservation of automatic and reflex motor activity — in the lower cranial nerve distributions (V, VII and IX through XII) of both sides. It was first described by Magnus in 1837 [8] and then reported almost exclusively in the French literature [1, 3, 5, 11] with some exceptions [2, 4, 9].

We report a typical case of FCM syndrome. It presented with an acute facio-pharyngo-glossomasticatory diplegia and automatic-voluntary dissociation of the affected muscles. CT-scan showed bilateral damage: the left hemisphere lesion was confined to the rolandic operculum; in the right hemisphere the whole territory of the middle cerebral artery was involved.

There are two main reasons for the present case report:

Firstly, even if FCM syndrome has a localizing value, it is often misdiagnosed because the neurological picture is very similar to that of the more frequent bulbar and peripheral palsies. Thus, from a clinico-methodological point of view, the procedures that led to the diagnosis in our case are worth emphasizing. Briefly: a) FCM syndrome, being the cortical type of pseudobulbar paralysis, can be differentiated from peripheral and bulbar syndromes on the basis of clinical and electromyographic signs of supranuclear neuronal damage; b) beyond the general characteristics of a pseudobulbar deficit, some peculiar clinical features of FCM syndrome point to a cortical localization of lesions; c) CT-scanning clinches the diagnosis by disclosing the bilateral perisylvian softenings including the rolandic operculum of both sides.

Secondly, the patient we describe was completely speechless, but not aphasic, as revealed by neuropsychological examination. Her speech

was totally disrupted while her ability to communicate was normal due to normal language organization. This dissociation is particularly interesting in the light of the CT-scans, because it confirms the hypotheses, recently advanced by some authors [6, 10, 12], that speech may be split into different components, which are differently localized in the left hemisphere. In summary: a) speech may be considered from two main points of view: the effectiveness with which information is transmitted (i.e.: 'communication') and the distinctness of enunciation (i.e.: 'articulation') [6]; b) impairment of communication and impairment of articulation are generally tightly interwoven in patients with anterior (pre-rolandic) left hemisphere damage

(Broca's aphasia); yet, as shown by a few cases reported in the literature [10, 12], they may be completely dissociated; c) there seems to be an anatomical basis for this dissociation: within the left hemisphere, lesions restricted to the foot of F3 (area 44 or Broca's area) transiently impair some 'linguistic' aspects of speech, but do not alter articulatory agility; by contrast, the destruction of the lower motor strip (with the rolandic operculum) exclusively affects articulatory abilities without involving language organization.

### Case report

This 47-year-old right-handed female is a dress-

*Fig. 1. Photographic record of the case, showing some of the clinical features described in the text. Apart from a congenital divergent strabismus, gaze to right (A), to left (B) and upward (C) are not defective. The facial-masticatory muscles and the tongue are motionless. Knitting and unknitting of the brows only appear as synkinetic movements in (A) and (C). The deficit of voluntary eyelid movements is documented in (D): the patient was asked to close her eyes; her unsuccessful attempts only resulted in knitting of the brows (synkinetic movement of the frontal musculature).*



maker. In May 1978 she sustained a moderate left facio-brachio-crural hemiparesis which cleared almost completely in two months. A mild articulatory difficulty was present in the acute stage for a few days. In the same year she was found to have a mitral stenosis and atrial fibrillation, previously unrecognized. In March 1977 she underwent valvular commissurotomy, after which prophylaxis with digitalis and anti-coagulants was started. The patient enjoyed relatively good health till February 1979, when she suddenly became totally unable to move her facial and masticatory muscles, to stick out her tongue, to swallow or to utter articulate sounds. Admission neurological examination revealed a facio-pharyngo-glosso-masticatory diplegia, a moderate motor and sensory deficit in the right arm, no visual field disturbances. In the following days the syndrome was examined in detail:

- The facial diplegia involved the lower and the upper portion of the VII nerve, yet without a Bell's sign. The facial musculature was motionless and hypotonic; the mouth was half-open (Fig. 1 A, B, C, D); voluntary eyelid movements could not be performed (Fig. 1 D), but the corneal reflex was spared and a brisk glabellar reflex could be elicited.

- Ocular movements were possible in all directions, but adduction of the left eye on gaze to the right appeared to be defective, owing to a congenital divergent strabismus (Fig. 1 A).

- Impairment of chewing, due to bilateral masticatory paralysis, was not associated with trigeminal sensory deficit. The masseteric reflex (jaw jerk) was exaggerated with a tendency to a clonic response.

- The tongue was completely immobile, flaccid, but not hypotrophic or fibrillating.

- The soft palate and fauces were also immobile. There was no "curtain-movement"; the palatal and pharyngeal reflexes (gag reflex) could not be elicited.

- Dysphagia was one of the most striking features, especially in the acute stage. The patient was unable to perform the first (voluntary) phase of deglutition, whereas the second (reflex) phase was intact. There was continuous drooling and saliva frequently ended in the trachea or in the nose.

- No paralysis of the vocal cords was found on laryngoscopy.

- Automatic movements were spared: the patient could knit and unknit her brows on looking upward (Fig. 1 C), blink, close her eyes while sleeping.

- Reflex movements such as coughing, sneezing, belching were also present.

- The patient could not communicate orally: she produced only inarticulate, unmodulated

sounds and answered questions only by moving her head and eyes or by writing.

*Electromyographic investigations*, performed with coaxial needle electrodes, on the orbicularis oculi, orbicularis oris and tongue muscles of both sides showed: no fibrillation at rest; complete absence of motor unit potentials during attempts at voluntary contractions whereas some motor unit potentials could be recorded from the orbicularis oculi during reflex and automatic movements of the eyelids.

The excitability of facial and hypoglossus nerves was normal and the evoked motor action potentials appeared to be normal in amplitude, form and latency.

Both early and late responses of the blink reflex, elicited with mechanical and electrical (0.5 msec duration, double threshold intensity) stimulations, were normal. The jaw reflex appeared after normal latency.

In conclusion: the electrophysiological study demonstrated lack of voluntary activity but no signs of peripheral motor neurone damage.

*CT-scan* showed two areas of decreased density: the left lesion destroyed the inferior portion of the rolandic cortex, partially extending deeply to the subcortical white matter; damage to the right hemisphere involved cortical, subcortical and deep structures of the frontal, parietal and temporal lobes (Fig. 2).

From the lateral reconstruction of CT maps (Fig. 3) it could be seen that the left lesion was circumscribed to the lower third of the frontoparietal gyri, including the rolandic operculum: the pars triangularis and the pars opercularis (area 44 or Broca's area) of the third frontal gyrus were spared. The right lesion extended over the whole territory of the middle cerebral artery.

#### *Neuropsychological Examination*

*Language*: speech was completely suppressed, the patient being unable even to vocalize. But her speech comprehension appeared to be intact. She scored 35/36 on a shortened version of the Token Test.

Phonemic, semantic and syntactic levels of language integration were explored for the receptive route, by means of three different tests:

- Semantic discrimination test (including subtests of auditory language comprehension and reading comprehension): the patient correctly selected, 39 out of 40 times, the picture corresponding to a word — read aloud by the examiner or printed on a sheet — among three pictured alternatives, strictly related from a semantic-conceptual point of view.

- Phoneme discrimination test: the patient could discriminate with 99% accuracy between two non-sense CCVC syllables pronounced aloud by the examiner.

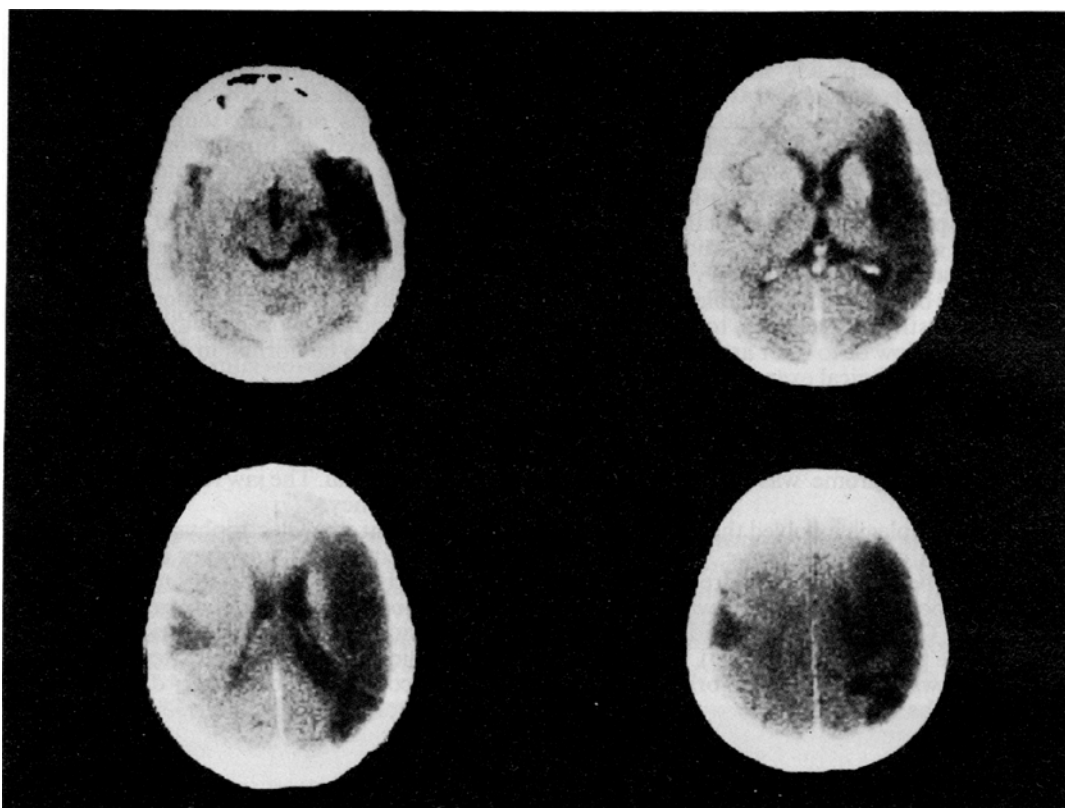


Fig. 2. CT-scan sections: two areas of decreased density are recognizable as ischemic lesions. The left lesion is confined to the rolandic operculum whereas the right lesion involves the whole territory of the middle cerebral artery.

—Syntax Battery: requested to point, out of 2 or 4 alternatives, to the picture representing a syntactically complex sentence, the patient showed normal comprehension of reversible sentences (active, passive, temporal relations — before/after —, embedded active and passive).

Spontaneous writing or writing about a subject set by the examiner was performed slowly but without phonemic and semantic paraphasias and with good syntactic construction. The patient was also capable of writing under dictation and of copying a block-letter text perfectly. Written naming of pictures was prompt and accurate.

**Memory:** digit-span (tested by pointing to the correct alternatives in a 10-choice device) was 6 forwards and 4 backwards. Spatial memory span (Corsi's Block Tapping Test) was 5 forwards and 4 backwards. Visual memory of meaningless shapes and of object drawings was normal (both immediate and delayed).

**Praxis:** the impairment of volitional movements did not allow testing for bucco-facial apraxia.

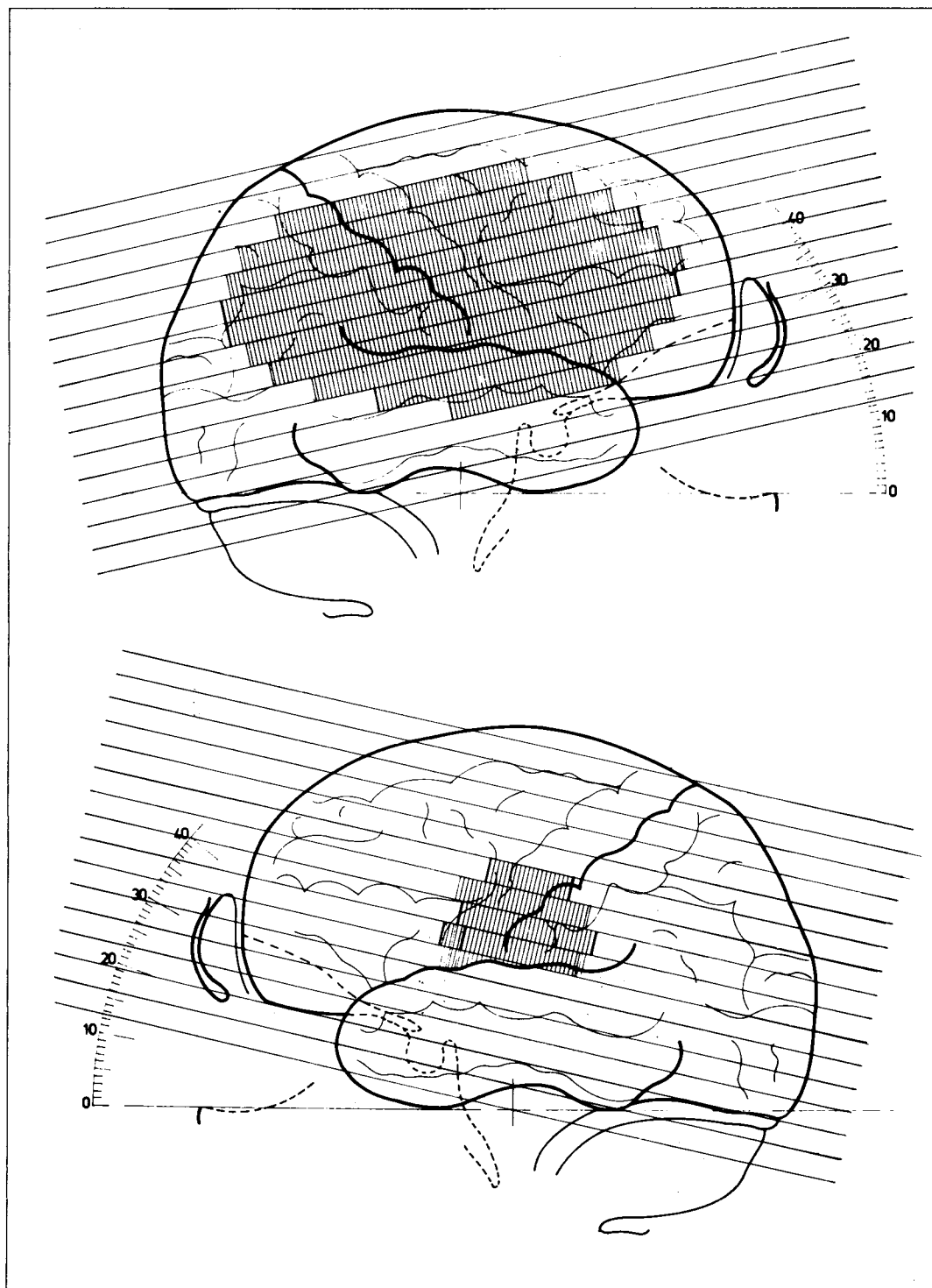
Transitive and intransitive gestures (including two and three element sequences), both on oral order and on imitation, were accurately performed with the right arm. More difficulty was noted for the left, owing to the slight pyramidal deficit. Constructional apraxia was absent, since the patient was capable of reproducing geometrical figures with a very good accuracy.

**Visuo-spatial abilities:** the patient had no difficulty finding her way about; could accurately copy geometric and figurative designs and arrange elements in their proper spatial relationship. Left-sided spatial neglect and other disturbances of visuo-spatial exploration of the extra-personal space (usually related to a massive right hemisphere damage) could not be evidenced.

**General intelligence:** In Raven's Coloured Matrices (1947), she gave 30 correct responses out of 36.

In conclusion: no neuropsychological deficit could be detected beyond the complete suppression of speech.

Fig. 3. Lateral reconstruction of the CT-scan sections shown in fig. 2. The precise boundaries of the infarctions in the cortex of right (top) and left (bottom) hemispheres are shown.



## Discussion

The clinical pattern was indicative of a pseudobulbar palsy, resulting from bilateral breakdown of the connexions between the cortical motor areas and the nuclei of the lower cranial nerves.

Peripheral palsy was ruled out by the absence of muscle denervation phenomena, such as fasciculation and hypotrophy, the presence of the blink reflex, the augmentation of the masseteric reflex, the sparing of some automatic motility. Likewise, bulbar damage (due to a vertebrobasilar ischemic stroke) would be incompatible with the lack of massive paralysis of the limbs, of sensory and internuclear signs. The electromyographic data, supplemented by blink and jaw reflex recordings, confirmed the clinical evidence of "upper" rather than "lower" motor neurone damage.

Other clinical features were then specific for a cortical localization of the lesions, thus suggesting the diagnosis of 'opercular syndrome', that is the cortical type of pseudobulbar paralysis:

- the association of lower cranial nerve involvement with a transient motor and sensory deficit of the right arm;

- the dissociation between the severe motor deficit and the absence of sensory symptoms in the territory innervated by the trigeminal nerve;

- the history of the patient, showing repeated CVAs with pyramidal motor weakness involving both sides alternately.

CT-scan supported a definite diagnosis of FCM syndrome by disclosing the cortical level of the bilateral lesions.

The second interesting feature in the present case is the dissociation between the normal language organization and the complete suppression of speech — that is, following Levine and Mohr (1979), between normal 'communication' and totally disrupted oral 'articulation' — and its CT-scan correlate.

It has recently been suggested [10, 12] that lesions confined to posterior F3 of the left hemisphere (area 44 or Broca's area) do not produce either Broca's aphasia or any persisting aphasic syndrome. They only give rise to mild, transient language disorders, word finding and naming difficulties, literal and verbal paraphasias, shor-

tening and simplification of sentences, moderate comprehension disturbances [12], without loss of articulatory agility.

On the other hand, selective damage to the left lower motor strip (including the rolandic operculum) only affects articulation causing a 'cortical dysarthria' (slowness and dysprosodic alteration of speech-melody with both stuttering and poor control of pitch [12]).

Our case lends further support to the notion that a left hemisphere lesion confined to the rolandic operculum and sparing Broca's area is not associated with a 'linguistic' impairment of speech, but only with a 'more peripheral' deficit of speech articulation. In our case the latter deficit was much more severe than the 'cortical dysarthria': since the left rolandic operculum lesion was superimposed on preexisting right hemisphere damage (including the right rolandic operculum), the typical clinical pattern of FCM syndrome (with complete inability even to vocalize) had developed.

The successive involvement of the right and left rolandic opercula by CVAs is also relevant to the notion of a different role of the fronto-parietal portions of the two hemispheres in speech. It has been claimed that fronto-parietal lesions of the non-dominant hemisphere, although massive, do not affect the efficiency of 'communication': they may impair 'articulation', but mildly and transiently. However, if a small fronto-parietal lesion of the left hemisphere — incapable in itself of producing aphasic disturbances or severe articulatory defects — follows this primary right hemisphere lesion, a very severe 'pseudobulbar dysarthria' takes place [6, 7]. On this interpretation, our patient's first stroke, though extensively involving the right hemisphere, only produced a very mild, transient dysarthria; it was the second, less severe, contralateral stroke (with an ischemic lesion circumscribed to the rolandic operculum) that caused suppression of speech, yet without aphasia.

A complete facio-pharyngo-glosso-masticatory diplegia, rather than a pseudobulbar dysarthria, occurred since the primary cortical motor areas for the lower cranial portions were destroyed in both hemispheres. Thus, the oft-reported dissociation between 'communicative' and 'articulatory' aspects of speech was displayed by our patient in the highest degree.

## Sommario

*Viene descritta una paziente portatrice di due lesioni ischemiche coinvolgenti l'opercolo rolandico in entrambi gli emisferi. Il quadro clinico era caratterizzato da una compromissione selettiva dei movimenti volontari (con integrità di quelli automatici e riflessi) bilateralmente a livello facio-faringo-glosso-*

masticatorio (sindrome di Foix-Chavany-Marie). La paziente, del tutto incapace di articolazione e fonazione, non risultava afasica ad una valutazione neuropsicologica. Questa dissociazione viene discussa in riferimento alla particolare localizzazione delle lesioni alla T.A.C. cerebrale.

Address reprint requests to: Dr. Giampiero Villa  
Clinica Neurologica  
Policlinico "A. Gemelli"  
Largo A. Gemelli, 8 — 00168 Roma

## References

- [1] ALAJOUANINE T. and THUREL R.: *La diplégie faciale cérébrale, forme corticale de la paralysie pseudobulbaire*. (Contribution à l'étude de la dissociation des activités volontaires et réflexes). *Rev. Neurol.* 40: 441-458, 1933.
- [2] BRUYN G.W. and GATHIER J.C.: *The operculum syndrome*. In: Vinken, P.J. and Bruyn, G.W. (eds.): *Handbook of clinical neurology*, Amsterdam, North Holland, 1969.
- [3] CHÂTEAU R., FAU R., GROSLAMBERT R., PERRET J., BOUCHARLAT J. and CHÂTELAIN R.: *A propos de trois observations de diplégie linguo-facio-masticatrice d'origine corticale: la forme de l'adulte et celle de l'enfant*. *Rev. Neurol.* 114: 390-395, 1966.
- [4] FERRARI G., BONINSEGNA C. and BELTRAMELLO A.: *Foix-Chavany syndrome: CT study and clinical report of three cases*. *Neuroradiology* 18: 41-42, 1979.
- [5] FOIX C., CHAVANY J.A. and MARIE J.: *Diplégie facio-linguo-masticatrice d'origine cortico-sous-cortical sans paralysie des membres*. *Rev. Neurol.*, 33: 214-219, 1926.
- [6] LEVINE D.N. and MOHR J.P.: *Language after bilateral cerebral infarctions: role of the minor hemisphere in speech*. *Neurology*, 29: 927-938, 1979.
- [7] LUDWIG M.E.: *Beitrag zur Frage der Bedeutung der Unterwerigen Hemisphäre*. *Z. Gesamte Neurol. Psychiat.* 164: 735-747, 1939.
- [8] MAGNUS A.: *Fall von Aufhebung des Willenseinflusses auf einige Hirnnerven*. *Müllers Arch. Anat. Physiol. Wissensch. Med.*, 258-266, 1837.
- [9] MARIANI C., SPINNLER H., STERZI R. and VALLAR G.: *Bilateral perisylvian softenings: bilateral anterior opercular syndrome (Foix-Chavany-Marie syndrome)*. *J. Neurol.*, 223: 269-284, 1980.
- [10] MOHR J.P., PESSIN M.S., FINKELSTEIN S., FUNKENSTEIN H.H., DUNCAN G.W., and DAVIS K.R.: *Broca aphasia: pathologic and clinical*. *Neurology* 28: 311-324, 1978.
- [11] SCHOTT B., BOULLIAT G., COTTE L., and VAUTERIN C.: *Le syndrome operculaire bilatéral et unilatéral*. *Lyon Med.* 206: 365-378, 1961.
- [12] TONKONOGY J. and GOODGLASS H.: *Language function, foot of the third frontal gyrus and Rolandic operculum*. *Arch. Neurol.* 38: 486-490, 1981.